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CASE REPORT

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Coronary Sinus Ostial Obstruction in Single-Ventricle Congenital Heart Disease

Two Patients With Different Outcomes

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ABSTRACT

Coronary sinus ostial obstruction is an exceedingly rare anomaly that is particularly important to diagnose in patients with single-ventricle heart disease before surgical palliation. We present 2 cases, an infant and an adult, diagnosed with coronary sinus ostial obstruction, with different clinical outcomes due to timing of diagnosis. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1459–1462) Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

Coronary sinus ostial obstruction (CSOO) with a persistent left superior vena cava (LSVC) is a rare anomaly with particularly important implications for patients with single-ventricle heart disease set to undergo palliative surgery via bidirectional superior cavopulmonary anastomosis (SCPA) and/or total cavopulmonary anastomosis (TCPA). Although in an otherwise structurally normal heart, this finding would be of no clinical consequence, failure to diagnose this in a patient with single-ventricle heart disease before palliative surgery can lead to cardiac dysfunction and death.

LEARNING OBJECTIVES

- To highlight the importance of CSOO in single-ventricle congenital heart disease.
- To emphasize the necessity of ruling out CSOO before single-ventricle palliative surgery.
- To present echocardiographic and angiographic findings in patients with CSOO.
- To suggest echocardiographic and catheter-based approaches for ruling out CSOO.

We present 2 cases of CSOO with persistent LSVC, one in an infant before surgical palliation and one in an adult with ongoing cardiac dysfunction after direct right atrial to pulmonary artery connection (classic Fontan).

CASE SERIES

PATIENT 1. A 3-month-old infant with a double inlet left ventricle, normally related great vessels, and an

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ABBREVIATIONS AND ACRONYMS

CS = coronary sinus

CSOO = coronary sinus ostial obstruction

LIV = left innominate vein

LSVC = left superior vena cava

SCPA = superior cavopulmonary anastomosis

TCPA = total cavopulmonary anastomosis

unrestrictive bulboventricular foramen presented to the cardiac catheterization laboratory for hemodynamic catheterization and evaluation of the branch pulmonary arteries before bidirectional SCPA.

The patient was prenatally diagnosed with the congenital cardiac defect and after birth underwent balloon atrial septostomy followed by main pulmonary artery banding at 10 days of life. At 3 months, routine echocardiography showing main pulmonary artery band encroachment on the left pulmonary artery prompted further evaluation by cardiac catheterization.

As part of pre-bidirectional SCPA catheterization, balloon occlusion angiography performed in the left innominate vein (LIV) revealed an LSVC to coronary sinus connection. There was no egress of contrast from the coronary sinus (CS) as expected; however, once the balloon in the innominate vein was deflated, there was retrograde drainage of contrast from the cardiac veins superiorly through the LSVC and into the LIV, consistent with a diagnosis of CSOO with persistent LSVC (**Figure 1A**, Video 1). After angiographic diagnosis, retrospective review of echocardiographic images with color and spectral Doppler showed abnormal superiorly directed flow in the LSVC, draining into the LIV (**Figure 2**).

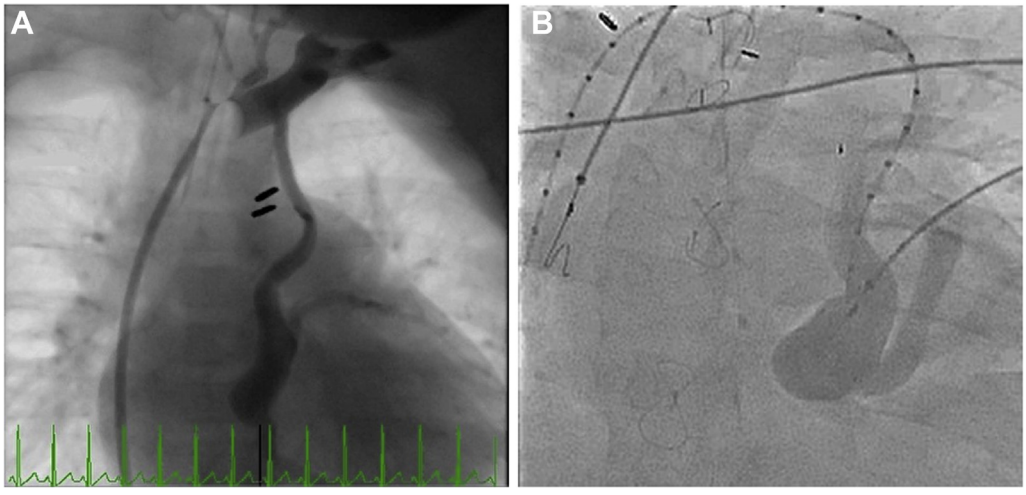
The patient underwent bidirectional SCPA; due to concern for impedance of coronary venous drainage if the CS was left connected to the SCPA circuit, unroofing of the CS was also performed by using a technique similar to that described by Ohta et al (1). The LSVC was divided distally, and a 2-mm coronary probe was inserted and tunneled inferiorly into the CS behind the left atrium. Through an atriotomy, an incision was made along the probe into the CS, thus providing an unobstructed opening for coronary venous egress (**Figure 3**). The LSVC was then ligated distally. Post-operative echocardiogram showed normal cardiac function with unobstructed flow from the CS into the left atrium. The patient had an uncomplicated post-operative course and was discharged home for continued routine follow up.

PATIENT 2. A 27-year-old man with tricuspid atresia presented to the catheterization laboratory for hemodynamic catheterization and electrophysiology study in anticipation of possible "Fontan revision" surgery.

The patient was postnatally diagnosed with tricuspid atresia with normally related great vessels, and he underwent atriopulmonary Fontan with left pulmonary arterioplasty at 1 year of age. There was immediate and persistent depressed cardiac function after the Fontan operation; over the course of the next 2 decades, the patient also experienced atrial arrhythmias, necessitating chronic medication use, transcatheter radiofrequency ablation, and multiple electrical cardioversions.

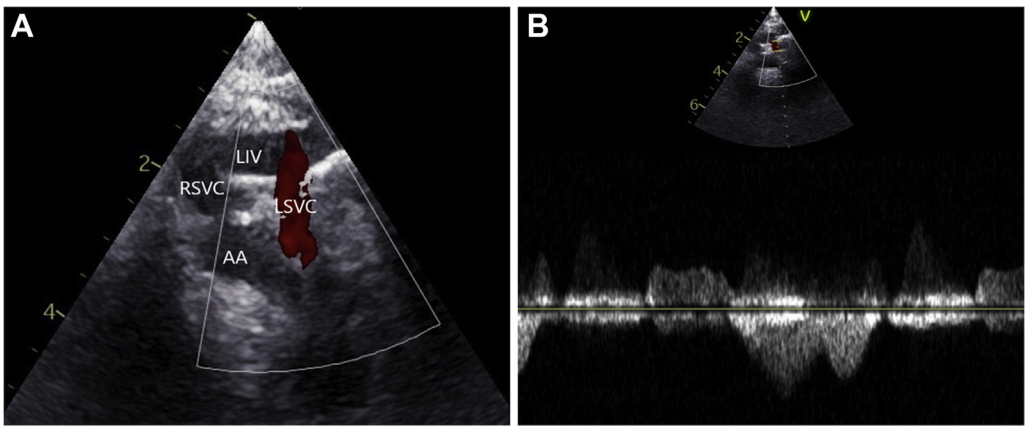
Due to his ongoing arrhythmias and cardiac dysfunction in the setting of an atriopulmonary Fontan, the patient was put forward for tomographic imaging, cardiac catheterization, and electrophysiology study to determine candidacy for classic Fontan

FIGURE 1 Left Superior Vena Cava Angiography



Left innominate vein **(A)** and left superior vena cava **(B)** angiography in an infant and adult, respectively, with coronary sinus ostial obstruction. The coronary sinuses are dilated with more pronounced dilatation of the cardiac veins in the adult.

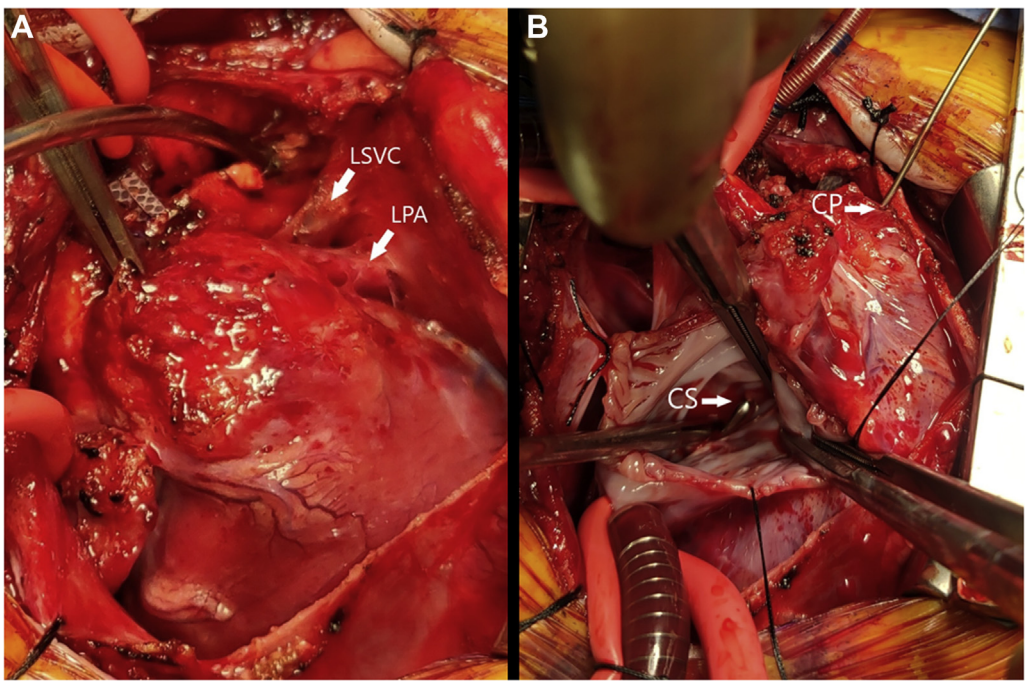
FIGURE 2 Echocardiography of Retrograde LSVC Flow



Color Doppler **(A)** and pulse wave spectral Doppler echocardiography **(B)** from a suprasternal view shows the superiorly directed flow in the left superior vena cava (LSVC), draining into the left innominate vein (LIV). AA = ascending aorta; RSVC = right superior vena cava.

revision to an extracardiac Fontan. Transesophageal echocardiogram revealed a dilated CS that was not previously seen by transthoracic echocardiogram due to poor acoustic windows, along with continued left ventricular dysfunction with a shortening fraction of 22%. Upon injection of contrast into the LIV, an LSVC to CS connection was noted with severe dilation of the CS and cardiac veins (**Figure 1B,** Video 2). There was CSOO with near atresia of the CS os that was also confirmed by cardiac computed tomography imaging.

FIGURE 3 Intraoperative Photographs of CS Unroofing



Intraoperative photographs showing the orientation of the left superior vena cava (LSVC) coursing posterior to the left pulmonary artery (LPA) **(A)** and a 2-mm coronary probe (CP) inserted into the LSVC and tunneled down to the surgically unroofed coronary sinus (CS) **(B)**.

Although the left ventricular diastolic function was preserved, the calculated cardiac output was mildly diminished, and there was evidence of pulmonary hypertension with atriopulmonary Fontan circuit pressures and transpulmonary gradient that were moderately elevated, making the patient a poor candidate for Fontan revision.

After a 6-month trial of vasodilator medication, the patient returned to the catheterization laboratory for repeat hemodynamic study, which revealed continued mildly diminished cardiac output with improvement in Fontan pressures and transpulmonary gradient. Thought was given toward performing a catheter-based intervention to relieve the obstruction, much like that performed by Petit et al (2); however, the patient was deemed a candidate for Fontan revision, and thus his CS will be unroofed at the time of surgery. Although there are a multitude of reasons for cardiac dysfunction after single-ventricle palliation, this patient's dysfunction began immediately following his palliative operation and has persisted, making the newly found CSOO a likely contributor to the ongoing dysfunction.

DISCUSSION

Diagnosis of CSOO and persistent LSVC is critical before bidirectional SCPA or TCPA in staged single-ventricle palliation. Most patients with a diagnosis of single-ventricle heart disease and bilateral superior vena cavae will have both caval veins divided and anastomosed to the pulmonary arteries; however, if the LSVC is particularly small and there is a connecting vein between the 2 cavae, the LSVC may simply be ligated to prevent venous runoff. If the LSVC is the only available route for cardiac venous egress, ligation would result in acute cardiac venous hypertension, myocardial ischemia, and possibly death, as seen in several previous case reports (2-6). If the LSVC is not recognized and/or is preserved, there will be hindrance of cardiac venous egress secondary to the higher pressure in the cavopulmonary system after SCPA or TCPA. This also will lead to diminished myocardial perfusion and cardiac dysfunction, as seen in one of our patients. Preoperative diagnosis will allow for proper surgical management and avoidance of potentially catastrophic consequences. Although surgical results of creating a CS to left atrial communication have been successful, there have also been successful cases of minimally invasive, transcatheter approaches to relief of CSOO (2,7,8).

Although diagnosis of CSOO with persistent LSVC can be challenging, multiple echocardiograms are typically performed on patients with single-ventricle congenital heart disease, which can offer clues to the diagnosis and prompt further investigation. For instance, the presence of an LSVC with retrograde flow and/or dilation of the CS, as seen in one of our patients, should clue the clinician in to the possible diagnosis. To that end, a thorough search for an LSVC should be performed via echocardiogram, along with spectral Doppler interrogation of the LSVC if found. Given the limitations of transthoracic echocardiography, especially in often unruly infants, selective angiography in the LIV by cardiac catheterization is advisable before SCPA or TCPA operations to rule out CSOO with persistent LSVC.

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KEY WORDS adults with congenital heart disease, children, congenital heart surgery, coronary sinus ostial atresia, coronary sinus obstruction, single-ventricle congenital heart disease

APPENDIX For supplemental videos, please see the online version of this paper.